Case Report

Primary T Cell lymphoma of Small Intestine with Perforation is a Sinister Combination: A Case Report of Two Cases

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Abstract

Introduction: Primary T cell lymphoma of the gastrointestinal tract is a relatively rare heterogeneous group of lymphoid malignancies comprising various subtypes. Perforation is a serious life threatening complication of gastrointestinal tract lymphoma. Usually, perforation occurs after initiation of chemotherapy but rarely, it may be the initial presentation of the gastrointestinal tract lymphoma. T cell lymphoma of the small bowel presenting or getting complicated by perforation is of grave prognosis.

Materials and Methods: We identified 2 cases of small intestine primary T cell lymphoma managed in a surgical unit of a tertiary care hospital. A retrospective review of data was done to elucidate clinical-pathological features and prognosis.

Results: Of the two patients identified, one was a 45 year old male presented with peritonitis secondary to perforation in the ileum. He underwent emergency laparotomy segmental enteral resection and anastomosis. The second patient was a 58 year old female patient admitted with complaint of recurrent abdominal pain. After surgical management and histopathological diagnosis, she was administered chemotherapy. She had perforation after the first cycle of chemotherapy. Both patients died 35 and 28 days respectively after first presentation to hospital.

Conclusion: Our report highlights the fact that intestinal T cell lymphoma complicated by perforation either at the time of presentation or during the course of chemotherapy is of grim prognosis.

Key words: T cell lymphoma, Intestinal lymphoma case report, Gastrointestinal tract, Perforation

Introduction

The gastrointestinal tract is the most common site of extranodal lymphomas and the majority of these are of B cell lineage [(1)]. Primary T cell lymphoma is relatively uncommon and constitutes 13-15% of primary gastrointestinal tract lymphoma in East

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Asia [1]. Primary T cell lymphoma is associated with poor prognosis and high mortality [2]. We present two cases of small intestine T cell lymphoma managed in a single surgery unit of a tertiary care institute. One case was presented with features of perforation peritonitis and another one presented with recurrent abdominal pain but complicated by perforation following the first cycle of chemotherapy. The following cases highlight the poor prognosis of small intestine T cell lymphoma developing perforation and the importance of understanding the histopathology and early start of treatment. The case report is in accordance with 2023 SCARE guidelines[3].

Presentation of the cases

Two cases of small intestine T cell lymphoma managed surgically in a surgical unit of tertiary care institute were identified. Retrospective analysis of the data was done. Clinical presentation, diagnostic workup, surgical management and details of chemotherapy were noted. Outcomes of therapy in both the patients were reviewed.

Case 1. A 45 year old male presented in emergency ward with a complaint of sudden onset of severe diffuse abdominal pain and vomiting for the last 12 hrs. He was a known case of right side hemiparesis secondary to cerebral thromboembolism for the last 4 years and was on phenytoin therapy for the same. He had a past history of intermittent abdominal pain for the last 2-3 months with associated weight loss. For these symptoms, he was evaluated elsewhere with CECT abdo66men which was suggestive of segmental thickening of the wall of ileum with associated lymphadenopathy (Fig 1). He had been on anti-tubercular therapy for one week. There was no history of celiac disease. On examination, the patient was malnourished and in distress due to severe abdominal pain. There was diffuse tenderness with voluntary guarding. Chest and abdomen X-ray showed free air under the diaphragm. Biochemical evaluation revealed anemia (Hb-7.7gm %) and leukocytosis (TLC-17800), rest of the investigations were with in normal limits. After evaluation and resuscitation, he underwent emergency laparotomy.

Per operative findings showed

a. Gross peritoneal contamination by purulent fluid.

b. About 65 cm proximal to ileocaecal junction, 8 cm long ileal segment was densely adherent to the ascending colon with a 2x2cm sized perforation in it.

c. About 120cm and 140 cm from the ileaocaecal junction respectively, 2 segments of ileum were inflamed with interloop and omental adhesions with the normal segment in between.

d. There were multiple enlarged jejunal and ileal mesentery lymph nodes.

The segmental resection of perforated ileum and ileoileal anastomosis was done.

On the third postoperative day, patient had hiccoughs, abdominal pain and low urine output. On examination, the abdomen was distended with a diffuse tympanic note. TLC (Total leukocyte count) was elevated i.e 21,600. He was managed conservatively with IV fluid, active nasogastric aspiration and antibiotic therapy. He responded to the above mentioned measures and started on a liquid diet after removing the nasogastric tube. However, on the sixth postoperative day, he complained of abdominal pain, vomiting and fever. TLC was persistently elevated i.e 23900. In view of recurrence of symptoms and raised TLC, CT scan of the abdomen was done to exclude anastomotic dehiscence and to detect any neo perforation in the inflamed segments of the ileum. The CT scan revealed features of small bowel obstruction with minimal degree of free fluid in the peritoneal cavity and residual free air. The patient responded to conservative management and was resumed on a semisolid diet. However TLC did not have any decreasing trends despite stepping up of antibiotic therapy. On resumption of the oral diet, the patient again had symptoms of abdominal distension, bilious vomiting and fever on postoperative day eleven. A CECT scan of the abdomen was done again. It showed irregular thickening of the wall of distal jejunum and proximal ileum with anexoenteric component leading to focal dilatation and narrowing of the lumen of the small bowel and enlarged mesenteric lymph nodes. TLC was persistently elevated, the rest of the biochemical examination was with in normal limits. He was managed conservatively with reinsertion of the

nasogastric tube, parenteral nutrition and intravenous fluids. He showed improvement with these measures and successfully resumed on a semisolid diet.

Histopathology and immunohistochemistry of the resected enteral segment showed infiltration of the small intestine with lymphoepithelial cells from small to intermediate size with irregular nuclei. The lymphoid cells were seen infiltrating into adjacent mesentery. Cells were positive for CD3/CD7/CD8/CD43/LCA. Occasional B cells were showing positive stains to CD 20. Cells were negative for CD4/CD2. The final impression was high grade T cell Non-Hodgkin's lymphoma (NHL) not otherwise specific.

In view of the recurrent attacks of sub-acute intestinal obstruction in the postoperative period early dose of chemotherapy was started with the objective to help in resolving these debilitating attacks. The first cycle of chemotherapy comprising Dexamethasone, Vincristine and cyclophosphamide was administered, which was well tolerated. However, before the next scheduled cycle of chemotherapy, the patient developed fever and succumbed to respiratory failure. His duration of survival from presentation to hospital to death was 35 days.

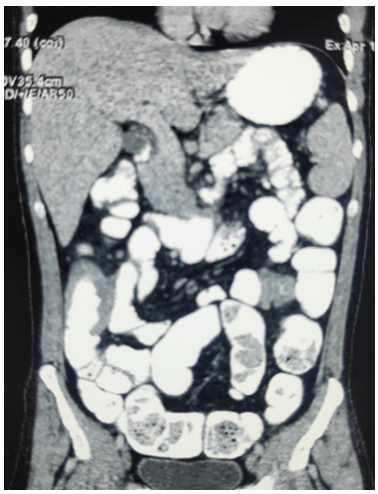

Fig 1. Thickened ileal loop

Case no 2- A 58 year old female presented in outpatient clinic with a six months history of recurrent diffuse abdominal pain for the last 6 months. She had a past history of appendectomy 8 years back. CECT scan abdomen showed segmental thickening in the jejunum/proximal ileum (Fig 2.). There was no history of celiac disease. Elective laparotomyrevealed

- Two non-obstructive strictures, 60cm and 75cm, for duodenojejunal flexure respectively.

- Multiple nodules in the serosa and mesentery adjacent to the stricture (Fig 3.).

A biopsy on mesenteric nodule was done. Her postoperative recovery was uneventful. Histopathology and immunohistochemistry showed peripheral T cell lymphoma high grade with Ki 67 of 80-85% positive for LCA (CD45)/CD 5/CD 7/CD 30/CD 45(Fig 4.). Tumor stained negative for CD2/CD3/Alk/CD20/Pax5/CD79a/Pancytokeratin. Chemotherapy comprising injections Cyclophosphamide, Dexomethasone, Vincristine and Adriamycin started. On the postchemotherapy dayone, patient developed severe diffuse abdominal pain. Abdominal ultrasound revealed free air with free intraperitoneal fluid. She was taken up for emergency laparotomy. Peroperative findings showed 4x4cm perforation 30 cm from duodenojejunal flexure, the perforated segment was adherent to adjacent jejunal loops and transverse mesocolon. Resection of the perforated segment followed by end to end anastomosis of ileum was done. In the postoperative period she had persistent hypotension and septicemia, non-responsive to resuscitative measures. She failed to respond and died of multi-organ failure on postoperative day 4. Her duration of survival from diagnosis was 28 days.

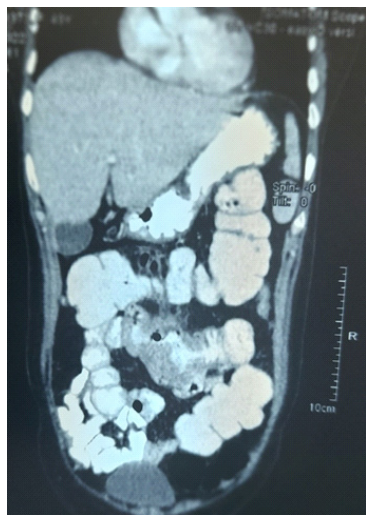

Fig 2. Thickened jejunal loop

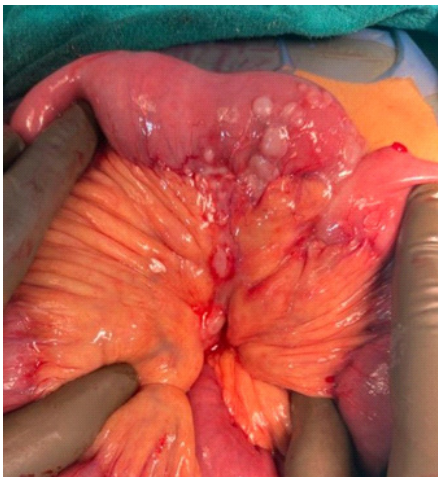

Fig 3. Stricture with multiple serosal & mesenteric nodules

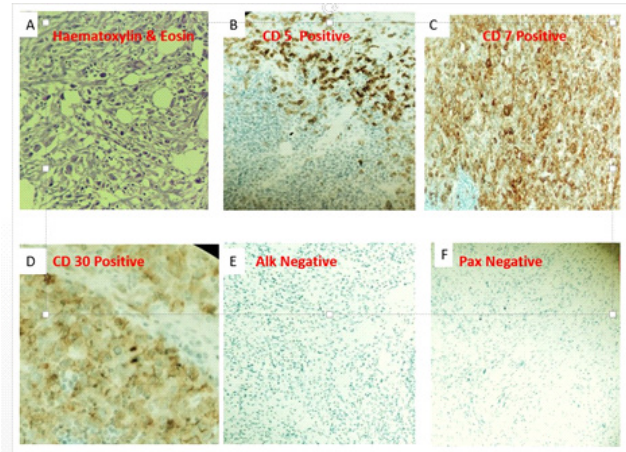

Fig 4. Haematoxylin eosin & Immunohistochemistry

Discussion

Primary intestinal lymphoma is a rare entity with a poor prognosis. It is predominantly found in the ileum (60-65%), followed by the jejunum (20-25%), duodenum (6-8%) and other sites (8-9%) [4] . Intestinal T cell lymphoma comprises four subtypes; 1. Enteropathy associated T-cell lymphoma (EATL), 2. Monomorphic and epitheliotropic intestinal T-cell lymphoma (MEITL), 3. Intestinal T-cell lymphoma not otherwise specified (NOS), 4.Provisional entity indolent T-cell lymphoproliferative disorder of the gastrointestinal tract (WHO classification 2017) [5]. There are no specific symptoms of intestinal lymphoma, majority of patients present with abdominal pain (70-80%), loss of weight (30%), G I bleeding (25.9%), diarrhea (16.9%) and rarely as perforation and intestinal obstruction [6]. Although primary GI lymphoma of the small bowel presenting as perforation is rare, it is not uncommon for the bowel to perforate post chemotherapy [7]. Preoperative diagnosis of T cell lymphoma is difficult, usually the diagnosis is made on the pathological examination of specimen.

We report two cases of primary intestinal T cell lymphoma. One was presented as a case of small bowel perforation and another presented with a complaint of abdominal pain but developed perforation after the first cycle of chemotherapy. Both of our patients had no history of celiac disease or any symptoms of malabsorption. Intestinal T cell lymphoma are clinically aggressive malignancies. They have worse 5 year survival compared to low grade B cell NHL at 23.8% versus 66.3%[8]. Both of our patients had a short duration of survival, 35 and 28 days respectively. This is in confirmation of a poor prognosis of gastrointestinal tract lymphoma complicated by perforation as reported in the literature [9]. Love J et al[10] reported a case of primary T cell lymphoma presenting as perforation peritonitis similar to our first case. In the postoperative period there patient had high ileostomy output requiring total parenteral nutrition. Tian Y et al[11] also reported a case of perforation peritonitis in 84 year old male secondary to primary T cell lymphoma. Their case had uneventful outcome and was able to complete adjuvant chemotherapy. Management of primary T cell NHL is multimodal, requiring both surgery and chemotherapy.

Conclusion

The clinical presentation of primary T cell lymphoma of a small bowel is non-specific. These cases emphasis the poor prognosis of small intestine primary T cell lymphoma associated with perforation. The non-specific clinical presentation and radiographic findings of primary T cell lymphoma reiterates the fact that histopathology with supportive immunohistochemistry is essential for accurate diagnosis. Multidisciplinary approach is required for adequate management.

Ethical Clearance: Institute ethical committee ref SGRD/IEC/2023-182, dated 19-01-2023

Conflict of interest: nil

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